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Presentation of a Hemangioblastoma in cavernous sinus: An extremely rare case report

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ABSTRACT

INTRODUCTION: Hemangioblastoma (HB) is a benign vascular tumor that accounts for about 2% of intracranial neoplasms. HB of the cavernous sinus (CS) is extremely rare. Only one report was found in the literature.

PRESENTATION OF CASE: We present a 29-year-old female with progressive headache and she had right ptosis and right mild oculomotor nerve palsy. The brain Magnetic Resonance Imaging (MRI) revealed a right extra-axial 4 × 4 cm in right CS position. The patient was operated upon microscopically via sub-temporal approach through a right temporal craniotomy. After the subtotal removal of a highly vascular tumor, the patient was referred for adjuvant therapy with Gamma-knife surgery.

DISCUSSION: Surgical removal of HB is the most effective treatment of the central nervous system (CNS) HBs. Tumors invading the CS could cause severe bleeding during surgery and HB—because of its vascular origin—had more risk for severe intraoperative bleeding, and in some cases resulted in surgery stop with subtotal resection of tumor.

CONCLUSION: It was recommended to perform a pre-operative brain angiography and selective embolization of these highly vascular lesions that it could result in subsequent complete surgical removal.

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1. Introduction

Hemangioblastoma (HB) is a benign vascular tumor that accounts for about 2% of intracranial neoplasms [1]. 33% of these tumors have the familial inheritance pattern and are linked to the von Hippel-Lindau (VHL) disease. The other 67% are sporadic [2]. HB is usually found in the infratentorial position and the supratentorial location is rare [3]. HB of the cavernous sinus (CS), as the patient of this study, is extremely rare and only one report was found in the literature [4]. This case is reported in line with the SCARE criteria [5].

2. Presentation of case

We present a 29-year-old female with progressive headache since one year. The patient's headaches were intermittent. After childbirth (cesarean section) six weeks ago, she suffered from continuous headache and double vision. She complained of nausea and vomiting and encountered right ptosis and right mild oculomotor nerve palsy without pupillary dysfunction on physical examina-

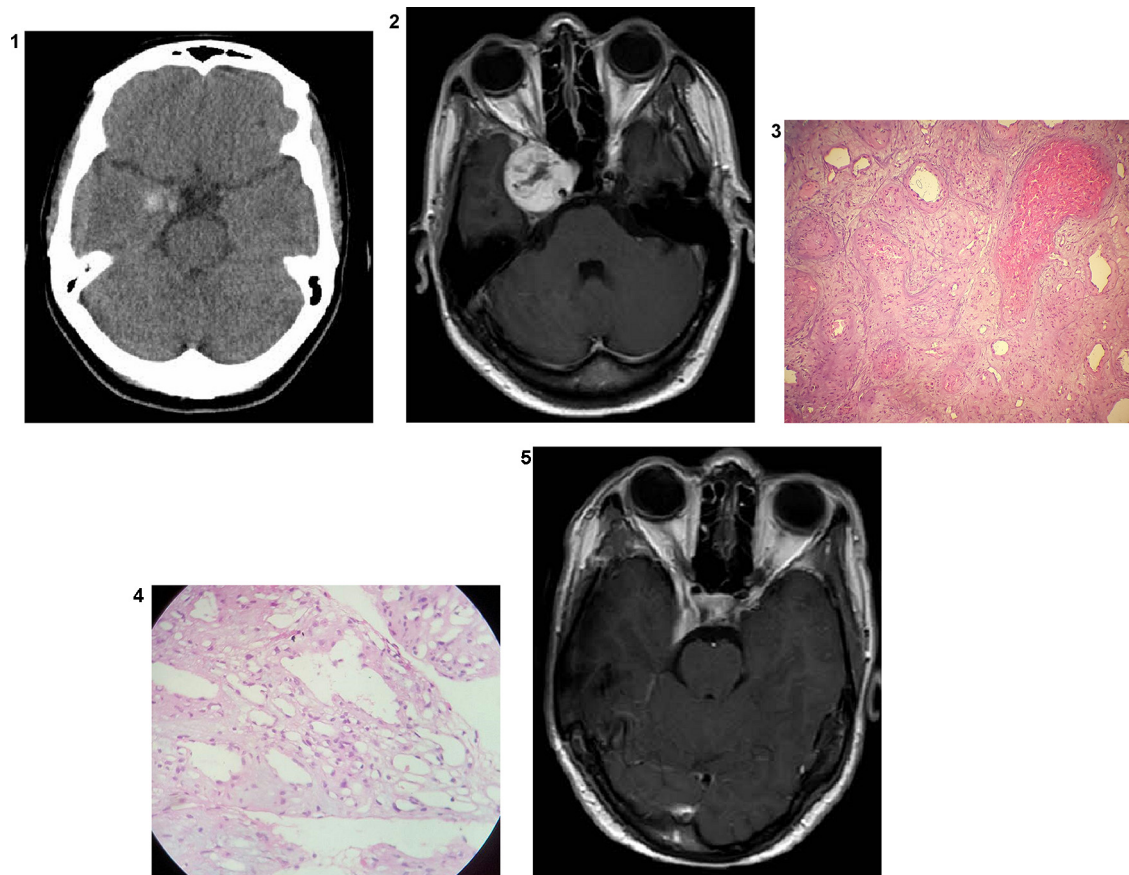
tion. On conducting a brain Computed tomography (CT) scan, a right medial temporal small high density (Fig. 1) was found. A brain Magnetic Resonance Imaging (MRI) was performed with and without Gadolinium (Gd) that revealed a right extra-axial 4 × 4 cm in the right CS position, with a bright enhancement that compressed the right medial temporal lobe (Fig. 2). Just one week after the patient's admission, she was operated microscopically via sub-temporal approach through a right temporal craniotomy in semi-lateral position. After subtotal removal of a highly vascular tumor, the surgery had to be stopped due to severe intraoperative bleeding and possibility of a cranial nerve palsy injury. In the post-operative period, the patient showed all signs and symptoms of improvement. The histopathologic report was Hemangioblastoma (HB) (Figs. 3 and 4). To assess the possibility of von Hippel-Lindau (VHL) disease; other complementary examinations such as ophthalmologic, renal and abdominal exams, and laboratory tests such as 24h-urine Vanillylmandelic Acid (VMA) was performed and none of these tests had any pathological finding. Four months later, the patient was followed-up upon and she again complained of double vision without any objective deficits. Another brain MRI was recommended with Gadolinium (Gd) that showed an enhanced remnant of the tumor on the right CS (Fig. 5). Therefore, the patient was referred the patient for adjuvant therapy with Gamma-knife surgery.

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Figs. 1–5. A preoperative brain CT scan (Fig. 1) revealed a right medial temporal small high density and brain MRI with Gd. Fig. 2 shows a well-defined 4 × 4 centimeter solid mass of right cavernous sinus with bright enhancing. Histopathologic study shows a highly vascular tissue with a predominant capillary portion. Fig. 3, H&E × 40). Between small vessels, numerous vacuolated stromal cells with foamy cytoplasm can be seen, containing lipids and hyper-chromatic pleomorphic nuclei (Fig. 4, H&E × 200). Finally, a postoperative MRI with Gd (Fig. 5), after 4 months, shows an enhancing remnant of tumor on right cavernous sinus.

3. Discussion

HB is a highly vascular tumor that is more prevalent in infratentorial locations, such as the cerebellum and spinal cord [3]. Two-thirds of these neoplasms are non-familial and sporadic. Only 30–33% of them are VHL-related and are familial tumors [2]. In the familial type, a neurosurgeon must investigate for concomitant pathology such as pheochromocytoma, renal cell carcinoma (RCC), renal cysts, pancreatic tumor and cyst, ependymal cyst adenoma, broad ligament cystadenoma, retinal HB, and finally endolymphatic sac tumors [6]. Now, high-risk patients for VHL undergo blood sample for testing VHL mutation. Our patient had no pathological finding on complementary tests.

Surgical removal of HB is the most effective treatment of the central nervous system (CNS) HBs. The HB in supratentorium is rare and only one report for cavernous sinus HB can be found. The CS has very important components such as, cranial nerves and carotid artery that increase complexity of surgical treatment of tumors of this area. Tumors invading the CS could cause severe bleeding during surgery and, because of its vascular origin, HB has more risk for severe intraoperative bleeding. During surgery, the patient faced a lot of bleeding that limited the surgical field and the surgery had to be stopped with subtotal resection of the tumor. Stereotactic Radiosurgery (SRS) such as Gamma-knife surgery can give a good rate of tumor control and improve neurological function in intracranial HB, especially for the sporadic type [7]. For this reason, the patient was referred for complementary SRS therapy.

Finally, as a helpful tool, a preoperative brain angiography and if possible, a preoperative selective embolization of these highly vas-

cular CNS tumors could help the neurosurgeons to conduct precise and complete surgical treatment for such patients.

4. Conclusion

We present an extremely rare case of cavernous sinus HB with complete explanation of its diagnosis process, surgical treatment, and follow-up that had good results. A preoperative brain angiography and selective embolization of these highly vascular lesions is recommended that can result in subsequent complete surgical removal. Stereotactic Radiosurgery (SRS) can provide a good rate of tumor control and improve neurological function in cases of subtotal resection of intracranial HB.

Conflicts of interest

The authors had no any conflicts of interest.

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Ethical approval

Our institution exempts ethical approval for case reports.

Consent

The authors had a written and signed consent to publish a case report from the patient.

Author contribution

Dr. Alireza Tabibkhoei: study concept or design.
Dr. Arash Fattahi: data interpretation, writing the paper.
Dr. Hessam Rahatlou: data collection.

Guarantor

Dr. Arash Fattahi is the Guarantor of this submitted manuscript.

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